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sc0035 194 Atrial and Ventricular Septal Defect Closure

Au16 *Todd Mendelson, MD, Carlos Alviar, MD, Muhamed Saric, MD, PhD*

s0170 ATRIAL SEPTAL DEFECT CLOSURE

p1245 Aside from the bicuspid aortic valve, an atrial septal defect (ASD) is the most common congenital cardiac anomaly in adults with an approximate prevalence of 1 per 1000 individuals.[1]This chapter focuses on echocardiographic imaging during ASD closure. Embryology, classification, diagnosis, and hemodynamic significance of ASDs are discussed in detail in the congenital heart disease section of this book. Briefly, there are four main types of ASDs (listed in decreasing order of frequency): secundum ASD, primum ASD, sinus venosus ASD, and unroofed coronary sinus. When indicated, ASDs can be closed either surgically or via a percutaneous approach. According to most recent guidelines,[2] the primary indication for ASD closure is the presence of right atrial or right ventricular enlargement regardless of symptoms. Closure of ASD may also be considered:

o0010 • In the setting of either paradoxical embolism or platypnea-orthodeoxia. Paradoxical embolism refers to an embolus originating in the venous circulation that crosses into the systemic circulation via a shunt (such as an ASD). Platypnea-orthodeoxia refers to a syndrome of arterial oxygen desaturation when changing from a recumbent to an upright position.

o0015 • In the presence of left-to-right shunt, pulmonary artery pressure less than two-thirds systemic pressure, pulmonary vascular resistance less than two-thirds systemic resistance, or when responsive to pulmonary vasodilators or occlusion test. During occlusion test, ASD is transiently closed using sizing balloon and the patient's hemodynamic parameters are monitored. Closure is aborted if hemodynamic instability or signs of acute pulmonary edema develop.[3]

p1260 The primary contraindication of ASD closure is the presence of irreversible, severe pulmonary arterial hypertension and no evidence of left-to-right shunt.

s0175 Surgical Atrial Septal Defect Closure

p1265 The earliest surgical closure of an atrial septal defect was reported in the early 1950s.[4] Surgical ASD closure was the first successful open heart operation (performed under general hypothermia and inflow occlusion) even before the advent of cardiopulmonary bypass.[5] Subsequently, ASD closure became the very first type of cardiac surgery to use cardiopulmonary bypass.[6] Surgical closure can be accomplished by either direct suture or using a patch. It is recommended that the surgical ASD closure be performed by surgeons with expertise and special training in congenital heart disease.[2] Surgery remains the only recommended means of closing primum, sinus venosus, and coronary sinus types of ASDs. Surgery is an alternative to percutaneous closure of secundum ASDs.

s0180 Percutaneous Atrial Septal Defect Closure

p1270 Percutaneous closure of an ASD was first described in the mid-1970s.[7] Currently, percutaneous closure has become the most common means of repairing secundum ASDs. All currently available ASD closure devices in the United States are only approved for secundum-type ASDs. These devices have a similar basic structure; they all contain two discs connected by a waist. Some are approved for simple secundum ASDs with a solitary hole, whereas others are specifically designed for secundum ASDs with multiple holes, referred to as fenestrated or cribriform (sievelike) ASDs. The three most commonly used devices (Fig. 194.1) are:

- Amplatzer atrial septal occluder (St Jude Medical, St Paul, Minn) o0020 is used to close nonfenestrated secundum ASDs. It contains a larger left atrial disc connected to a smaller right atrial disc. The waist connecting the two discs ranges from 4 mm to 38 mm in diameter. When selecting appropriate device size, the waist diameter of the devices should correspond to the ASD diameter.
- Gore Helex atrial septal occluder (W. L. Gore & Associates, o0025 Flagstaff, Ariz) contains two equal-sized discs connected by a spiral shaft; disc diameter ranges from 15 to 35 mm. An appropriately selected Gore Helex device should have a disc diameter that at is at least twice the ASD diameter.
- Amplatzer multifenestrated atrial septal occluder (St Jude Med- o0030 ical) contains two equal-sized discs connected by a thin shaft for use with cribriform ASDs. Disk diameters range from 18 to 35 mm. An appropriately selected device should have a disc size of a sufficient diameter to cover the entire ASD.

Typically, the percutaneous approach is used to close simple p1290 secundum ASDs, that is, those not associated with other congenital anomalies that might need surgical repair.

Role of Echocardiography in Percutaneous Atrial Septal Defect Closure s0185

Echocardiography is an essential part of the percutaneous ASD clo- p1295 sure process, as it is needed before, during, and after percutaneous ASD closure.

Before Atrial Septal Defect Closure s0190

Both transthoracic and transesophageal echocardiography can p1300 establish the presence of an ASD, define its type, size the defect, and determine shunt direction and its hemodynamic significance. Three-dimensional (3D) echocardiography overcomes many limitations of two-dimensional (2D) echocardiography by providing accurate visualization of the size and shape of the defect and its rims on unique en face views. Proper ASD sizing is essential in selecting the device size to avoid complications from using an undersized or oversized closure device (such as incomplete defect closure, device embolization, or disc erosion into surrounding cardiac structures).

In general, when deciding on the size of an ASD closure device, p1305 the maximum diameter of a secundum ASD cannot exceed the device specific cutoff value, which is 38 mm when using an Amplatzer atrial septal occluder and 18 mm when using a Gore Helex device. Furthermore, there should be sufficient ASD rims to anchor the device. Anatomically, there are six distinct ASD rims listed in a clockwise direction: superior vena cava rim, aortic (anterior) rim, atrioventricular rim, inferior vena cava (IVC) rim, posteroinferior rim, and posterosuperior rim.[8,9]

Historically, the device size was selected based on an invasive p1310 measurement of a so-called stop-flow ASD diameter by gradually inflating a sizing balloon placed across an ASD until no color Doppler flow across the ASD is seen on transesophageal echocardiography (TEE). More recently, device selection is based on direct ASD diameter measurements by 2D and 3D TEE.

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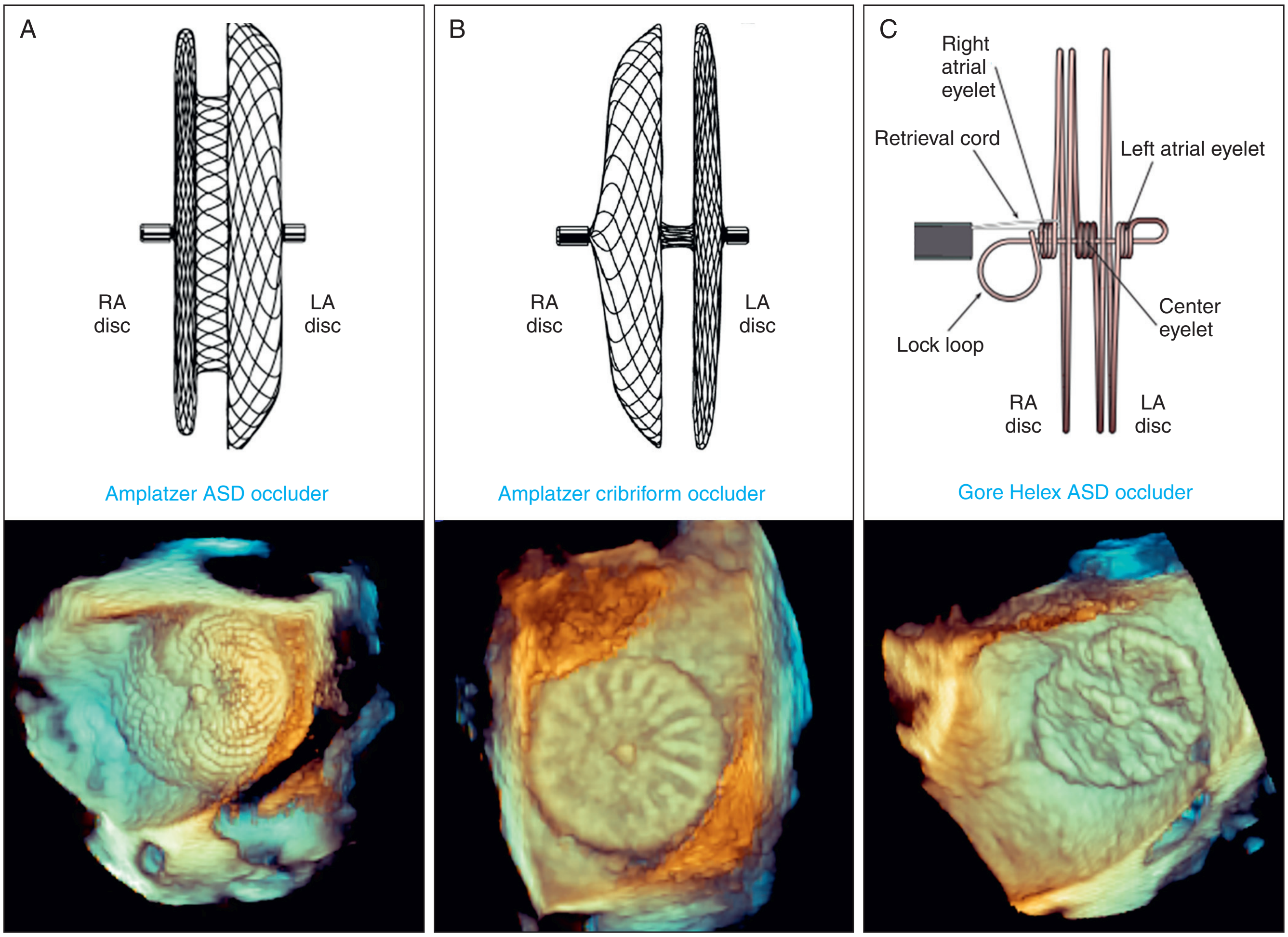

f0110 **Figure 194.1.** Secundum atrial septal defect (ASD) occluders. The three devices most commonly used for percutaneous ASD closure are depicted. *Bottom* image in each panel represents an en face three-dimensional transesophageal echocardiographic zoom view of the left atrial disc. *LA,* Left atrium; *RA,* right atrium.

p1315 On 2D TEE, ASD should be visualized in multiple views; in each view, the maximum ASD diameter during atrial diastole as well as the size of the two visible ASD rims should be measured. At approximately 0 degrees (four-chamber view), atrioventricular and posterosuperior ASD rims are seen. At approximately 60 degrees (short-axis view at the level of the aortic valve), aortic and posteroinferior rim are seen. At 90 to 120 degrees (bicaval view), superior vena cava (SVC) and IVC rims are visualized (Fig. 194.2/Video 194.2, *A* through *D*).

p1320 The term "sufficient rim" denotes a minimum rim width capable of securely anchoring the closure device. For the Amplatzer atrial septal occluder, the rims should be at least 5 mm; for the Amplatzer multifenestrated atrial septal occluder, the SVC and the aortic rim should be at least 9 mm. Absence of the IVC rim is considered a contraindication for device closure of a secundum ASD. Absence of the aortic rim is a major risk factor for device erosion into surrounding structure, especially when using the Amplatzer atrial septal occluder.[10]

p1325 3D TEE is especially well suited for accurate characterization of ASD size, shape, and rim (Fig. 194.3). On 3D zoom en face views, the full extent of ASD and its relations to surrounding cardiac structures are demonstrated from both the right atrial and left atrial perspectives. The so-called TUPLE (tilt up then left) maneuver can be utilized to place ASD images in anatomically correct orientation, which then facilitates characterization of ASD anatomy.[11] On 3D TEE imaging, one can easily determine the size of a secundum ASD (circular, ovoid, or irregular), its location within the floor of the fossa ovalis, and the presence of absence of associated anomalies such as an atrial septal aneurysm involving the remainder of the fossa ovalis.[12]

During Atrial Septal Defect Closure s0195

During the procedure, venous access is gained via the femoral vein. Subsequently, the interventionalist may opt to advance a sizing balloon across the ASD to confirm the ASD size using the stop-flow technique described earlier (Fig. 194.4, *A* and *B*). Thereafter, a delivery catheter is advanced into the right atrium and then through the ASD into the left atrium under fluoroscopic and echocardiographic guidance. A collapsed ASD closure device attached to its delivery cable is advanced through the delivery catheter, and the left atrial disc is opened first and positioned against the left atrial side of ASD. In the next step, the right atrial disc is opened to anchor the device within the ASD (see Fig. 194.4, *C* and *D*). p1330

2D and 3D TEE imaging, or alternatively intracardiac echocardiography (ICE), is used to ascertain proper positioning of the closure device. On 3D TEE, the near-field left atrial disc is easier to visualize than the far-field right atrial disc. Once the proper positioning of the ASD closure device is determined, the device is unscrewed from its delivery cable and released. p1335

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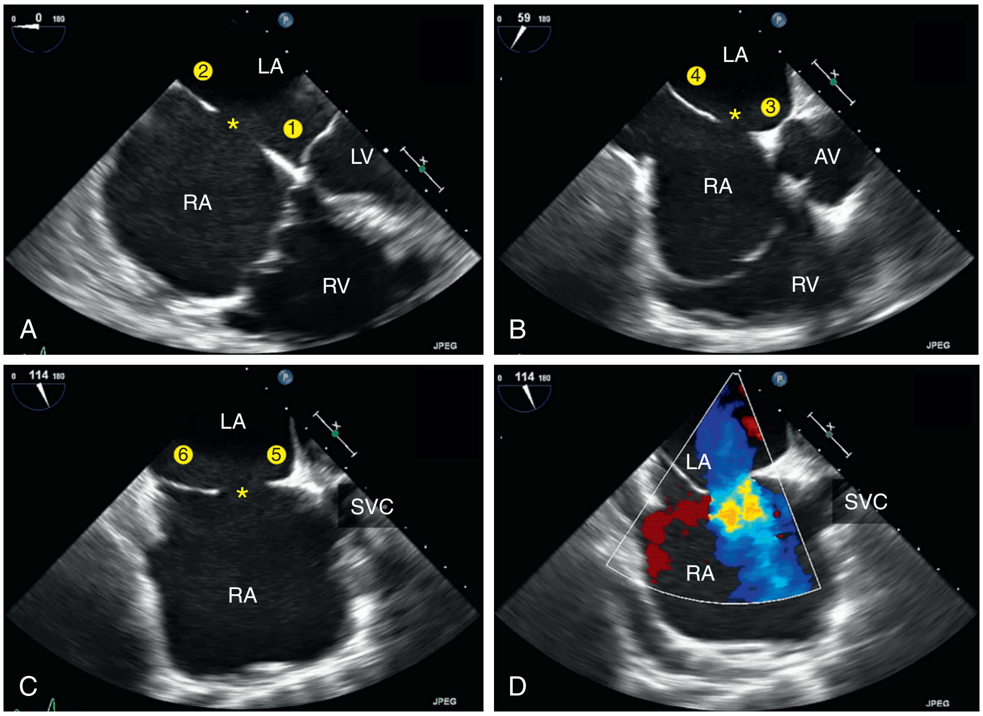
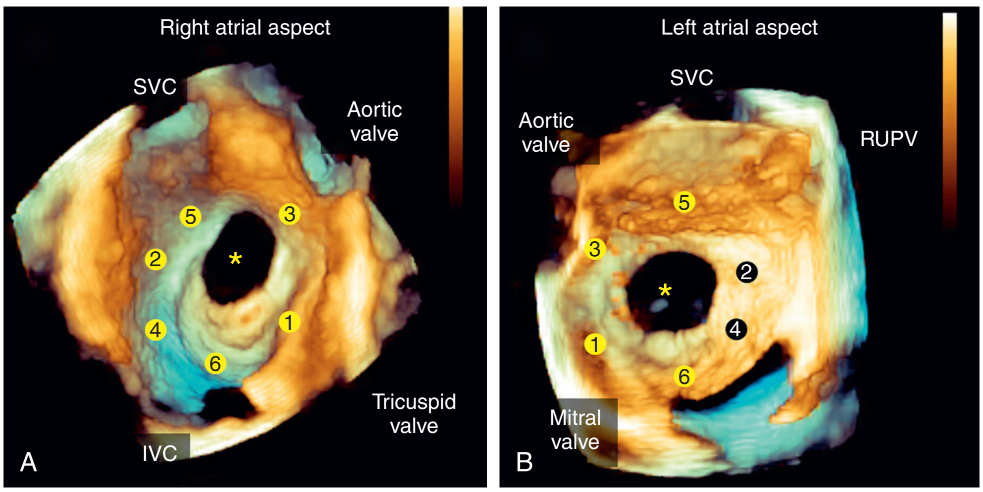


f0115 **Figure 194.2.** Secundum atrial septal defect (ASD) and its rims on two-dimensional transesophageal echocardiography. **A,** Secundum ASD *(asterisk)* seen at 0 degrees in the mid-esophageal 4-chamber view. Atrioventricular (❶) and posterosuperior (❷) ASD rims are seen. **B,** Secundum ASD *(asterisk)* seen at 59 degrees in the mid-esophageal short-axis view at the level of the aortic valve. Aortic (❸) and posteroinferior (❹) ASD rims are seen. **C,** Secundum ASD *(asterisk)* seen at 114 degrees in the mid-esophageal bicaval view. SVC (❺) and IVC (❻) ASD rims are seen. **D,** Color Doppler imagining at 114 degrees in the mid-esophageal bicaval view demonstrates a left-to-right shunt across the secundum ASD (from the left atrium to the right atrium). *AV,* Aortic valve; *IVC,* inferior vena cava; *LA,* left atrium; *LV,* left ventricle; *RA,* right atrium; *RV,* right ventricle; *SVC,* superior vena cava. (See
Au1 accompanying Video 194.2, *A* through *D*.)



f0120 **Figure 194.3.** Secundum atrial septal defect (ASD) and its rims on three-dimensional transesophageal echocardiography (3D TEE). 3D TEE zoom images of a secundum atrial septal defect *(asterisk)* from the right atrial (**A**) and left atrial (**B**) perspective. ASD is located in the anterosuperior portion of the fossa ovalis. The remainder of fossa ovalis is aneurysmal. ASD rims are clearly seen: ❶ Atrioventricular rim, ❷ Posterosuperior rim, ❸ Aortic rim, ❹ Posteroinferior rim, ❺ SVC rim, ❻ IVC rim. On these 3D TEE images, ASD was placed in proper anatomic orientation using the so-called TUPLE maneuver, which is demonstrated in the accompanying Video 194.3. *IVC,* Inferior vena cava; *RUPV,* right upper pulmonary vein; *SVC,* superior vena cava.

s0200 After Atrial Septal Defect Closure

p1340 Immediately after device release, 2D and 3D TEE is employed to check for device position, residual shunt, and presence of any complications such as a pericardial effusion.

p1345 When ASD closure is successful, color Doppler imaging demonstrates complete absence of any flow around the device (no peri-device leak between the edges of the closure device and ASD rims; see Fig. 194.4, *E*). In contrast, small amounts of color Doppler flow through the device are normal; they typically

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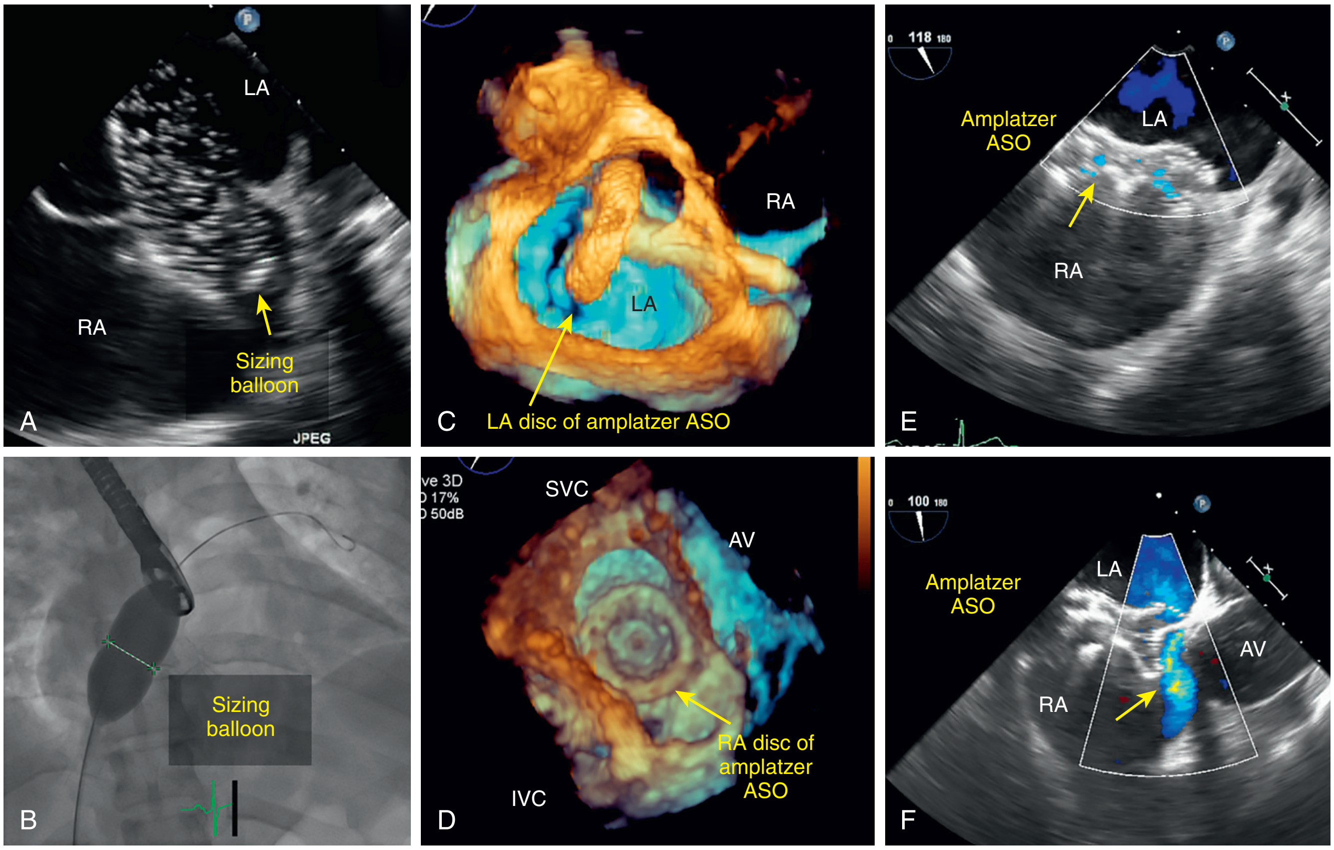

f0125 **Figure 194.4.** Percutaneous atrial septal defect (ASD) closure. Balloon sizing of a secundum ASD seen on two-dimensional transesophageal echocardiography (**A**) and fluoroscopy (**B**). Video 195-4, *A*, demonstrates that the balloon is inflated enough to prevent any shunt across the ASD. The diameter of the balloon at that moment is referred to as the stop-flow diameter. Deployment of an Amplatzer atrial septal occluder: first the left atrial disc is opened (**C**), followed by deployment of the right atrial disc (**D**). Post device deployment, color Doppler is used to assess proper closure. **E,** Successful ASD closure with only small amount of flow *(arrow)* through the device. Video 194.4, *E*, corresponds to this panel. **F,** Incomplete ASD closure with abnormal flow *(arrow)* around the device (para-device leak). Video 194.4, *F*, corresponds to this panel.

resolve over time as the device endothelializes (see Fig. 194.4, *F*). On completion of percutaneous ASD closure, the patient is placed on antiplatelet therapy for several weeks. Regular follow-up after closure, typically with transthoracic echocardiography, is recommended to ensure the absence of device migration, erosion, or other complications.

s0205 VENTRICULAR SEPTAL DEFECT CLOSURE

p1350 Ventricular septal defects (VSDs) can be divided according to their etiology as either congenital or acquired.[13] Acquired VSDs are less common than the congenital ones and are typically due to myocardial infarction or trauma.[14,15] VSDs can also be classified according to their anatomic location in perimembranous (also known as infracristal or subaortic), muscular (which can be subdivided into inlet, trabecular and infundibular, or supracristal), and atrioventricular or Gerbode defect, which entails a communication between the left ventricle and the right atrium.[16]

p1355 Perimembranous VSDs represent the majority of cases in post-neonates; they usually have a windsock appearance due to evagination of the membranous septum.[17] Muscular VSDs may be either acquired or congenital,[18] and they can be either solitary or multiple (when they may be referred to as “Swiss cheese VSDs”). When indicated, VSDs are typically closed surgically, although percutaneous closure options are being developed. According to current guidelines,[19] primary indications to close a VSD are:

- A pulmonary (Qp) to systemic (Qs) blood flow ratio (Qp/Qs) of 2.0 or greater AND clinical evidence of volume overload of the left ventricle o0035
- History of VSD-related infective endocarditis o0040

Closure of a VSD may also be considered in the following circumstances: p1370

- Qp/Qs ratio is greater than 1.5 AND pulmonary artery pressure is less than two thirds of systemic pressure AND pulmonary vascular resistance is less than two thirds of systemic vascular resistance o0045
- A net left-to-right shunting with a Qp/Qs ratio greater than 1.5 in the presence of left ventricular systolic or diastolic failure o0050

VSD closure is contraindicated in patients with severe irreversible pulmonary arterial hypertension. p1385

Surgical Ventricular Septal Defect Closure s0210

Surgery has been the classic approach to close VSDs. Steady improvements in surgical techniques have led to remarkable improvements in the prognosis and survival of patients with VSDs in the past 50 years.[20] However, a surgical VSD closure remains a major procedure requiring cardiopulmonary bypass and carries significant risk to the patient. Such risks are particularly high in patients with post–myocardial infarction VSDs who are frequently hemodynamically unstable and whose VSD borders are friable and difficult to suture.[21] p1390

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s0215 Percutaneous Ventricular Septal Defect Closure

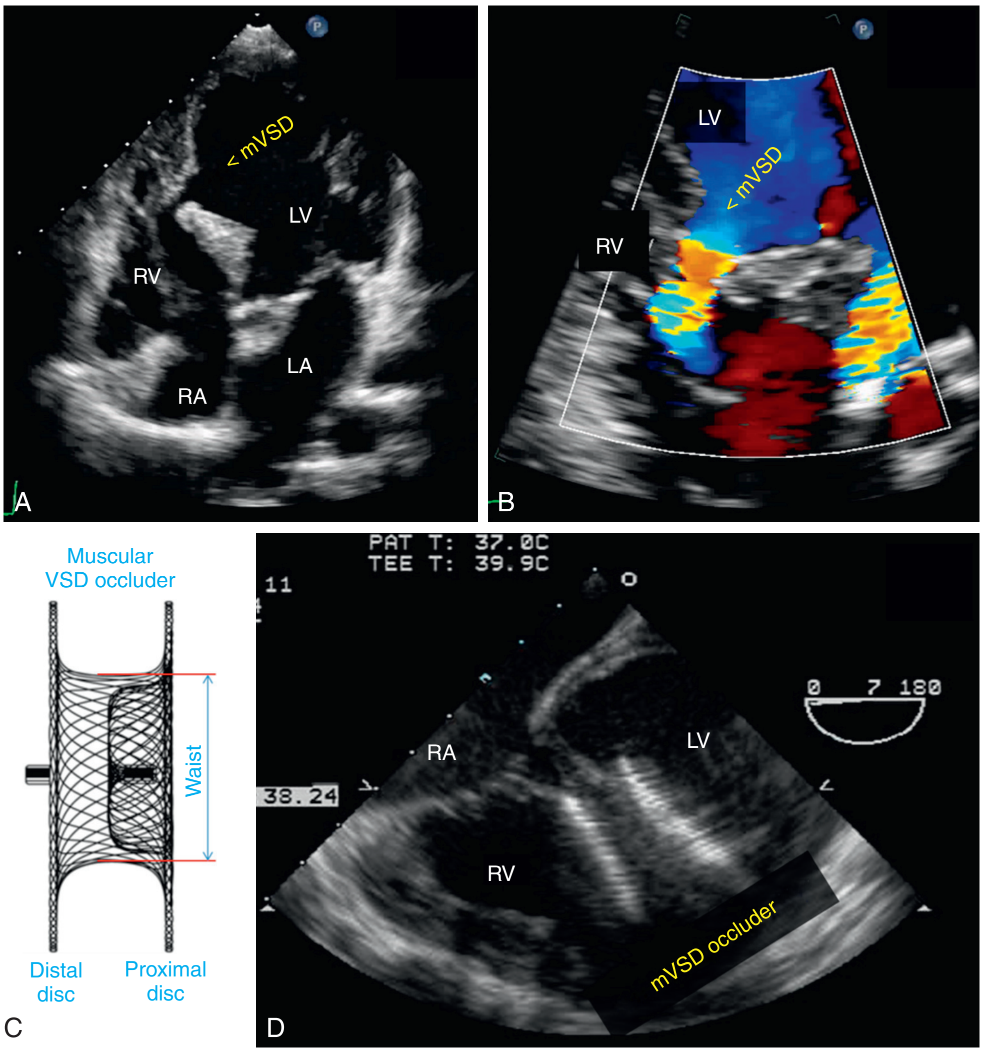
p1395 In the recent years, the use of percutaneous catheter-based devices has emerged as a nonsurgical option to treat VSDs in selected patients.[22] The first case of percutaneous VSD closure was reported in 1988 using a double-umbrella device.[23] Devices are currently approved in the United States for percutaneous closures of congenital VSDs that are not located in the proximity of heart valves in patients with high risk for surgical VSD closure.

p1400 Thus, congenital muscular VSDs are the principal VSD type amenable to percutaneous closure (Fig. 194.5). Postinfarction muscular VSDs have been closed percutaneously in an off-label manner.[24] In addition, percutaneous closure devices have been used to close residual ventricular defects after prior attempts at surgical closure as well as for traumatic or iatrogenic defects occurring after surgical aortic valve replacement.[25,26] Notably, because of their anatomic proximity to the heart valve, perimembranous VSDs and VSD associated with aortic valve prolapse are generally not amenable to transcatheter device closure unless surgical intervention is contraindicated.[27]

Different types of percutaneous devices have been tried for VSD closure; some of them yielded disappointing results, including the Rashkind double umbrella, the Bard Clamshell, the Button device and the Gianturco coils.[28] Currently, the Amplatzer Muscular VSD Occluder (St Jude Medical) is the only device specifically approved for VSD closure in the United States. It features two discs of equal diameter separated by a waist that is positioned across the VSD. It comes in different sizes with a width diameter ranging from 14 to 18 mm. An Amplatzer device with eccentric disc conformation specifically designed for closure of perimembranous VSDs (Fig. 194.6) has been used outside the United States.[29] p1405

Role of Echocardiography in Percutaneous Ventricular Septal Defect Closure s0220

Echocardiography plays an important role before, during, and after percutaneous VSD closure. Before closure, both transthoracic and transesophageal echocardiography can establish the presence of a VSD, define its type, size the defect, and determine its p1410



f0130 **Figure 194.5.** Congenital muscular ventricular septal defect (VSD). **A,** Transthoracic echocardiogram in the apical four-chamber view demonstrates a congenital muscular VSD *(mVSD)*. Note that the contractility of the surrounding interventricular septum is normal in congenital muscular VSD. This is in contrast to postinfarction muscular VSD, which is located within an area of interventricular septal hypokinesis or akinesis. **B,** Transthoracic echocardiogram with color Doppler in the apical four-chamber view demonstrates a left-to-right shunt across a congenital muscular VSD. **C,** Amplatzer muscular VSD occluder device approved in the United States for closure of congenital muscular VSDs. Note two symmetrical discs separated by a waist. **D,** Transesophageal echocardiogram in the four-chamber mid-esophageal view demonstrates an Amplatzer muscular VSD occluder. See accompanying Video 194.5, *A*, *B*, and *D*. *LA,* Left atrium; *LV,* left ventricle; *RA,* right atrium; *RV,* right ventricle.

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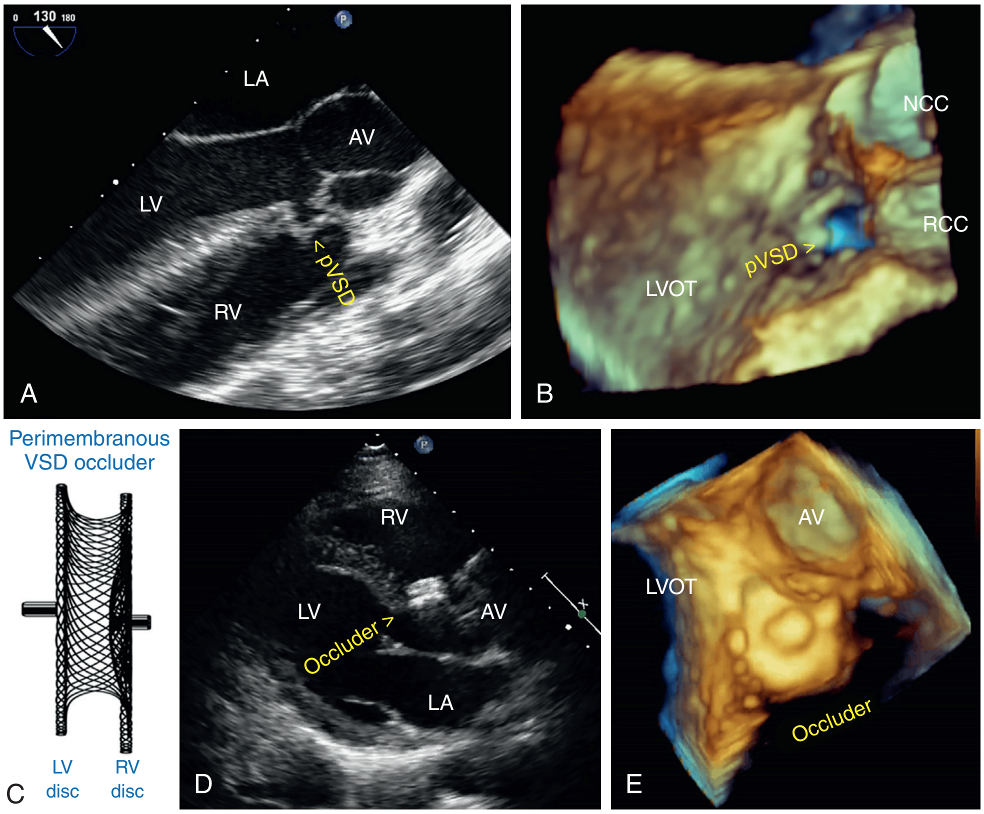

f0135 **Figure 194.6.** Congenital perimembranous ventricular septal defect (VSD). **A,** Transesophageal echocardiogram in a mid-esophageal view at 130 degrees demonstrates a windsock appearance of a congenital perimembranous VSD *(pVSD)*. **B,** Three-dimensional transesophageal echocardiography (3D TEE) zoom view demonstrates an en face view of a congenital perimembranous VSD just below the right coronary cusp *(RCC)* of the aortic valve. **C,** Amplatzer perimembranous VSD occluder device approved outside the United States for closure of congenital perimembranous VSDs. Note two asymmetrical discs separated by a waist. **D,** Transthoracic echocardiogram in the parasternal long-axis view demonstrates an Amplatzer perimembranous VSD occluder. Note in the video clip that there is no color flow around or across the device. **E,** 3D TEE zoom view demonstrates an en face view of an Amplatzer perimembranous VSD occluder. The device was implanted outside the United States. See accompanying Video 194.6, *A*, *D*, and *E*. Abbreviations: *AV,* Aortic valve; *LA,* left atrium; *LV,* left ventricle; *LVOT,* left ventricular outflow tract; *NCC,* noncoronary cusp of aortic valve; *RV,* right ventricle.

hemodynamic significance. 3D echocardiography may overcome limitations of 2D echocardiography[30] by providing accurate visualization of the size and shape of the defect on unique en face views of a VSD.[31,32] Proper VSD sizing is essential in selecting the device size to avoid complications from using an undersized or oversized closure device (such as incomplete defect closure or complete heart block).

p1415 Intraprocedural TEE, along with fluoroscopy, is essential for percutaneous VSD closure. 2D and 3D TEE is crucial for visualization of catheters, wires, and devices as they are being deployed in the heart. In general, percutaneous VSD closure is a complex procedure requiring both arterial and venous access. After arterial puncture, a catheter is delivered to the left ventricle in a retrograde fashion passing through the aortic valve, and its tip is placed across the VSD. Subsequently, using techniques of guide-wire snaring and exteriorizing to form an arteriovenous loop, an antegrade device delivery sheath is brought to the heart via the inferior vena cava after a venous puncture. The closure device is then advanced and carefully positioned across the VSD from the right ventricular side. The distal disc of the closure device is opened first and located on the left ventricular aspect of the VSD; this is followed by the deployment of the proximal disc on the right ventricular side.[33]

p1420 After percutaneous VSD closure, color Doppler imaging in conjunction with 2D and 3D imaging is essential for evaluating procedural success and possible complications. Successful VSD closure is characterized by a complete absence of any peri-device leak (a flow around the device between the VSD rims and the edge of the device). In contrast, small amounts of color Doppler flow through the device are normal and will resolve as the device endothelializes over time.

Please access ExpertConsult to view the corresponding videos for this chapter. p1425

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sc0040 195 Transcatheter Cardiac Pseudoaneurysm Closure

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p1430 Left ventricular pseudoaneurysm is a rare but serious complication of myocardial infarction, cardiac surgery, trauma, and infection. Medical treatment alone is frequently not effective and is associated with as much as 50% mortality. Until recently, the recommended treatment was surgical closure. These surgeries carried high risk due to abnormal hemodynamics, necrotic substrates, and the comorbidities of these patients.

p1435 Recently, transcatheter closure has been shown to be an acceptable alternative to open surgical intervention. Multimodality imaging, including three-dimensional (3D) echocardiography, identifies the location, size, and shape of the defect and can assess, guide, and follow up the closure procedure. This chapter discusses the use of transcatheter procedures in the treatment of an important complication of acute myocardial infarction, namely, left ventricular pseudoaneurysm.

p1440 Medical treatment of these patients carries very poor outcomes with high mortality rates. Surgery can close the defect but still involves high mortality rates. Transcatheter closure of these conditions is feasible and may be an alternative therapy in those patients with hemodynamic instability and other comorbidities. Real-time 3D echocardiography is an important imaging modality in the diagnosis and the assessment of this structural heart disease, and has a significant role in guiding and monitoring the interventional procedure.

LEFT VENTRICULAR PSEUDOANEURYSM s0225

Left ventricular free wall rupture is the most common acute tear of the left ventricle in patients with acute myocardial infarction. p1445 Unfortunately, in most patients acute free wall tear leads to severe intrapericardial bleeding, which rapidly results in cardiac tamponade and death. Therefore, although free-wall tear accounts for 14% to 20% of all myocardial infarction–related deaths, it is seen in only 7% of in-hospital myocardial infarction–related deaths.[1] In other words, most cases of death from acute free wall rupture occur before arrival in the hospital.

On rare occasions, the rupture is contained by pericardial and p1450 fibrous tissue, creating a left ventricular pseudoaneurysm. Characteristically, the orifice of the pseudoaneurysm is narrow, with a characteristic to-and-fro blood flow: from the left ventricle into the pseudoaneurysm during systole, and from the pseudoaneurysm into the left ventricle during diastole. The pseudoaneurysm wall is made of adherent pericardial or fibrous tissue, without any myocardial or endocardial layers. Thus, this wall is thin and may easily

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